Rare disease

Müllerianosis: a rare cause of acute renal colic

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Summary

A 50-year-old woman presented to the urology department with an acute history suggestive of left-sided renal colic. There were no other associated symptoms, but urine dipstick revealed non-visible haematuria. CT-KUB revealed a soft tissue mass at the left vesico-ureteric junction. Flexible cystoscopy demonstrated a mass intruding into the posterior bladder. A transurethral resection of the bladder 'tumour' was undertaken, and it was noted that the mass was not macroscopically consistent with transitional cell carcinoma. Histology demonstrated Müllerianosis, a rare lesion characterised by locally invasive growth of tissue originating from the Müllerian (paramesonephric) duct. The patient was seen by gynaecologist who initiated hormone treatment with an luteinising hormone - releasing hormone (LH-RH) analogue. Urological follow-up 3 months later highlighted ongoing pelvic pain but no further colicky loin pain. Repeat cystoscopy showed the mass had become smaller and the left ureter was laterally displaced. Further gynaecological input is planned if symptoms are ongoing.

BACKGROUND

Renal colic in adults is commonly due to renal calculi. Although initial investigations must always be conducted to exclude abdominal aortic aneurysm and urinary tract infection, the majority of cases are down to migrating renal stones. In this case, symptoms were caused by a bladder mass at the vesico-ureteric junction. Although initially thought to be transitional cell carcinoma, it was later identified as Müllerianosis, a rare lesion characterised by the proliferation of tissues derived from the embryological Müllerian duct. In this case, a common presentation strongly consistent with renal colic was found to be due to a rare condition.

Owing to its rarity, optimum management of this condition is currently unclear. Medical as well as surgical treatment approaches have been explored. Management in this case has encompassed both approaches as well as cross-departmental involvement.

Not only does this case demonstrate an alternative diagnosis in a case of renal colic, but it also highlights a rare, benign, alternative diagnosis for bladder masses, especially in women.

CASE PRESENTATION

We present the case of a 50-year-old woman presenting to acute surgery with a 3-day history of severe left-sided colicky pain. The patient had no urinary symptoms consistent with urinary tract infection. Her medical history included asthma and a long-term gynaecological history including dysmenorrhoea, menorrhagia and pelvic pain. The patient had had a Mirena coil inserted in 2008 but due to persistent symptoms underwent a laparoscopic subtotal hysterectomy. Histology undertaken following hysterectomy showed leiomyomas and endometritis. There was no relevant family or social history. Physical examination was unremarkable.

INVESTIGATIONS

Routine blood results were within normal limits. Urine dipstick demonstrated non-visible haematuria and urine cytology was negative for malignant cells. CT of kidneys, ureters and bladder (CT-KUB) demonstrated a 2 cm soft tissue lesion at the left vesico-ureteric junction with a normal, undilated left ureter. Further investigations included a CT urogram and a flexible cystoscopy which demonstrated a 2–3 cm area of raised urothelium suspicious of transitional cell carcinoma (TCC) (figure 1).

DIFFERENTIAL DIAGNOSIS

- Transitional cell carcinoma of the bladder
- Benign polyps of the bladder

TREATMENT

Transurethral resection of bladder tumour (TURBT) was undertaken. A mass that originated at the trigone was resected with loop diathermy. The lesion was not macroscopically consistent with a transitional cell carcinoma of the bladder and on excision produced a 'chocolatey exudate'. The bulk of the mass removed although some tumour remained.

OUTCOME AND FOLLOW-UP

Histology of the TURBT specimen demonstrated multiple glands throughout the lamina propria and muscularis layers. These included endosalpingioid, as well as endometrial type glands, with surrounding CD10 immunopositive endometrial stroma (figures 2 and 3). A diagnosis of Müllerianosis was made on the basis of the presence of these two types of glands of Müllerian origin.

Given the findings, both gynaecology and urology services were actively involved in follow-up. LH–RH analogues were initiated and a repeat cystoscopy was undertaken 3 months later. Symptoms of pelvic pain remained, although slightly improved and 'renal colic' was no longer a concern. Flexible

Figure 1 CT urogram demonstrating a mass protruding into the bladder and a non-dilated left ureter.

cystoscopy demonstrated residual, but much reduced, bladder mass with overlying urothelium. The left ureter was deviated postoperatively but a renal ultrasound scan is planned to ensure that there is no renal obstruction.

DISCUSSION

The purpose of this case is to raise awareness of a non-malignant cause of a symptomatic bladder mass, which more commonly presents in women. As this is a rare entity, the approach to and management of the condition is not yet clinically unified. In addition, our case demonstrates a rare presentation of a case of Müllerianosis.

Müllerianosis of the urinary bladder was first described by Young in 1996 as a condition whereby two of three Müllerian-derived tissues are identified within the bladder epithelium. These tissues are normal endometrium, endosalpinx or endocervix.¹ Müllerianosis is a rare condition with around 30 cases described in the literature to date. Most commonly, it has been identified in the urinary bladder but there are cases of it in other areas including abdominal scars² inguinal lymph nodes³ and other regions of the urinary tract.^{4–6}

Figure 2 Lamina propria of the bladder containing endometrial (A) and endosalpingiotic glands (B) (H&E $\times 4$).

Figure 3 Immunostaining showing CD10-positive stroma around endometrial glands (Immunohistochemistry $\times 10$).

In premenopausal women, commonly reported symptoms of pelvic pain, haematuria and filling-type lower urinary tract symptoms may be cyclical in response to hormonal changes. Nonetheless, case reports of older, symptomatic postmenopausal patients are also published.⁷ In such cases, basic first-line investigations to rule out urinary tract infection would be prudent. Depending on the age of the patient, further investigation with flexible cystoscopy and CT urography may be warranted in the absence of infection. In this case, it was prudent to eliminate renal stones, while in other cases presenting with pelvic pain, more gynaecologically focused investigations may be indicated. Colicky loin to groin pain may occur in the absence of proximal hydroureter and hydronephrosis.

The presentation of Müllerianosis in the urinary bladder has, thus far, only been highlighted in women. The distinctive location of the lesion, commonly on the posterior wall, has led to two hypotheses about its aetiology. The most widely accepted theory is of metaplastic origin although this fails to explain why the condition has not yet been described in the male urinary bladder.⁸ The alternative explanation is of direct implantation of cells during gynaecological instrumentation.^{7–9} In this case, surgical implantation is plausible; however, we feel that metaplastic change is the more robust and likely aetiology, as cases where no instrumentation has occurred have been reported.¹⁰

Macroscopically, areas of Müllerianosis have been identified to look very different to that of transitional cell carcinoma, and this may allow surgical suspicion of the histology to be raised at an early stage. It is easily differentiated from TCC on histology, with two of three Müllerian-derived tissue types being identified; endocervicosis, endosalpingiosis and/or endometriosis. These are usually identified as endometrial epithelium and stroma in endometriosis, tubal-type epithelium without surrounding stroma in endosalpingiosis and mucinous epithelium in endocervicosis. Using different staining techniques to identify the Müllerian cells can further cement the diagnosis; however, the tissues are distinctive in comparison with bladder epithelium and further stains may not be required. The cells are often seen in the muscularis propria giving it an invasive appearance; however, there is only one case in the literature that has demonstrated

malignant change in the urinary bladder and as such it is essentially considered to be a benign lesion.¹¹

Given that the Müllerian tissues are hormone responsive, efforts have been made to treat symptoms with hormone therapies, reducing hormonal stimulation and as such, reducing associated symptoms. Some available case reports have demonstrated both a reduction in symptoms and lesion size after a 3–6 month period of hormonal augmentation.^{7–10} Alternative approaches have been used, with one case report of a patient undergoing total abdominal hysterectomy and partial cystectomy to good effect.¹² Hormonal therapy alone has not been widely discussed. This is likely to be because a surgical approach to obtain a tissue diagnosis has usually been undertaken. The most common approach has been both surgical (transurethral resection of bladder tumour) and medical. Watchful waiting may also play a role in those with minor symptoms.⁷

Although this case report addresses a rare lesion, it highlights the importance of identifying appropriate and effective long-term treatment. In many cases the presence of Müllerianosis in the urinary bladder has led to unpleasant symptoms, causing significant morbidity to the patients involved. However, it is key to be aware that both medical options (hormonal agents) and surgical approaches (transurethral resection of bladder tumour and cystectomy) also carry significant morbidity. Although rare, a unified approach and understanding of the treatment of these lesions would allow more accurate recognition and focused treatment. It appears that Müllerianosis is a benign entity, with minimal malignant potential. Despite this, it appears that disease recurrence is common, and as such, optimum management may be similar to that of superficial bladder cancer, with regular surveillance and TURBT when symptomatic.

Learning points

- ▶ Renal colic in adults is common but bladder mass can be an unusual cause of this presentation.
- ▶ In women, Müllerianosis can be the cause of a symptomatic bladder mass. Both upper and lower urinary tract symptoms can be seen in these cases.
- ▶ Müllerianosis is a rare cause of urinary symptoms, especially in women but the condition, although infiltrating on histological specimens, runs a benign course.
- ▶ Combined surgical and medical intervention may be the best management option; however, given limited cases, optimal management strategies are yet to be identified.

Competing interests None.

Patient consent Obtained.

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